International Journal of Clinical Medicine, **2013, 4, 1-4**
Published Online December 2013 (http://www.scirp.org/journal/ijcm)
http://dx.doi.org/10.4236/ijcm.2013.412A1001

IgG4-Related Disease Presenting as a Soft Tissue Tumor Affecting Skeletal Muscle: A Case Report

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Received October 30th, 2013; revised November 28th, 2013; accepted December 15th, 2013

ABSTRACT

Background: IgG4-related disease is a systemic lymphoproliferative syndrome that shows IgG4-producing plasma cell expansion in affected organs with fibrotic or sclerotic changes. The lacrimal glands, salivary glands and pancreas are typically affected. We report a case of IgG4-related disease presenting a soft tissue tumor affecting skeletal muscle. **Case Report:** A 32-year-old man presented a soft tissue mass in his left arm. Magnetic resonance imaging revealed a spindle like, peripheral mass, in the lateral head of the triceps of his left arm. Tru-Cut Biopsy provided the diagnosis of IgG4-related disease affecting skeletal muscle. Glucocorticoid treatment was effective. **Conclusion:** To our knowledge, this is the first reported case of IgG4-related disease affecting skeletal muscle and presenting a soft tissue mass.

Keywords: IgG4-Related Disease; Soft Tissue Mass; Skeletal Muscle

1. Introduction

Immunoglobulin G4-related disease (IgG4-RD) is an increasingly recognized lymphoproliferative syndrome of unknown etiology. Patients show tumor-like swelling of involved organs, lymphoplasmacytic and IgG4 producing plasma cells infiltrating and affected organs with fibrotic or sclerotic changes. Elevated serum concentrations of IgG4 are found in 60% of patients with IgG4-RD.

The lacrimal glands, salivary glands and pancreas are the major affected organs.

Another feature of IgG4-RD is good glucocorticoid responsiveness.

We report the first case to our knowledge of IgG4-RD presenting a soft tissue tumor affecting skeletal muscle in the arm.

2. Case Report

A 32-year-old man was referred to our institution presenting a soft tissue mass in his left arm.

He had a medical history of asthma, left hypertrophic pachymeningitis and an orbital pseudotumor waiting for surgery.

Physical examination found a hard, nontender and mobile mass, with well defined borders, in the lateral aspect of his left arm, measuring 3 × 3 cm in size. It showed mild growing during the last two years. Tinel Test was negative. He also presented a right exophthalmos.

Complete blood count and serum chemistry, including IgG levels (787 mg/dl), were all within their normal limits (**Table 1**).

The MRI described a spindle like, peripheral mass, in the lateral head of the triceps, with well demarcated borders, T1 and T2 hyperintense and hypercaptant with contrast. It had a central hypointense region in all sequences that could correspond to fibrosis. It measured 2 cm × 17 mm × 2.5 cm (**Figure 1**).

An ultrasound guided Tru-Cut Biopsy was performed, referring 4 cylinders of material to Soft Tissue Pathology Department. The biopsy showed a fibro inflammatory condition, with abundant plasma cells, dissecting and

Table 1. Regular laboratory results.

Parameter	Value	Units
Erythrocytes (RBC)	4.21	$^{*}10^{6}/mm^{3}$
Hematocrit	43.2	%
White blood cell count	9.60	$^{*}10^{3}/mm^{3}$
Segmented neutrophils	78.7	%
Lymphocytes	9.9	%
Monocytes	9.5	%
Eosinophils	1.2	%
Basophils	0.7	%
INR	1	
Platelet count	163	$^{*}10^{3}/mm^{3}$
Fibrinogen	416	mg/dl
Creatinine, serum	1.04	mg/dl
Gamma glutamyl transferase (GGT)	17	U/l
Glucose	99	mg/dl
Lactate dehydrogenase serum (LDH)	4	mEq/l
Potassium	140	mEq/l
Sodium serum	74	mg/dl
Triglyceride	5.3	mg/dl
Uric acid		
Erythrocyte sedimenttion rate	2	mm/h
IgG serum	787	mg/dl
IgA serum	92.8	mg/dl
IgM serum	51	mg/dl

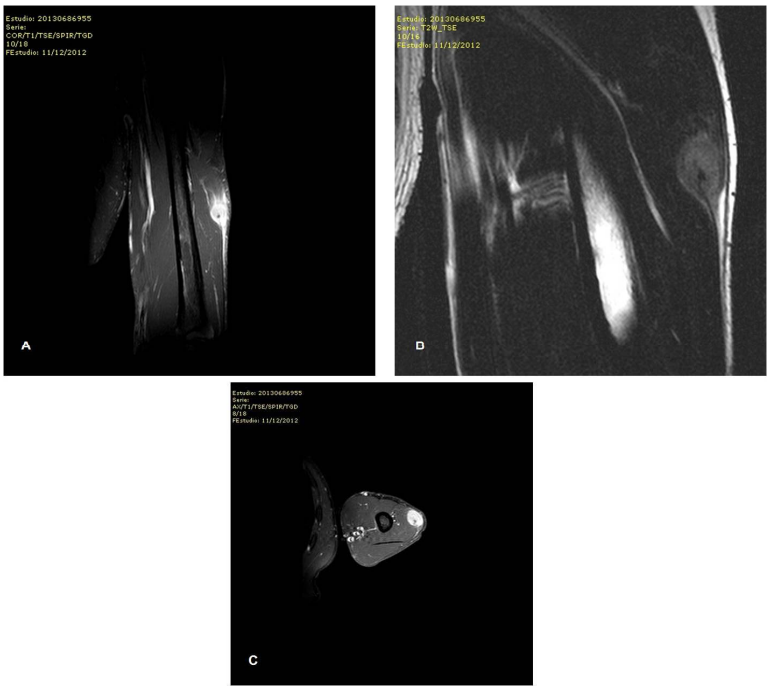

Figure 1. (A)-(C) Magnetic resonance images showed a well defined peripheral mass in the lateral head of the triceps hyper intense and hypercaptant with contrast.

infiltrating the skeletal muscle, causing degeneration and atrophy of muscular cells. Immunohistochemical analysis using CD 38 marker (plasma cell marker) and IgG4 stain revealed an increased IgG4 (+) plasma cell count (**Figure 2**).

Surgical treatment of his orbital pseudotumor was done. The biopsy of the orbital lesion showed an inflammatory pseudotumor. The soft tissue mass in his arm had a very good responsiveness to glucocorticoid treatment initiated after the neurosurgical procedure, reducing its size so no surgical treatment of the arm mass was indicated.

Based on the histopathological and immunohistochemical findings a diagnosis of IgG4-related disease affecting skeletal muscle was made.

3. Discussion

Soft tissue tumors are heterogeneous disorders, being the vast majority benign. They are most commonly characterized histologically according to the type of tissue they resemble. Unfortunately both benign soft tissue tumors and soft tissue sarcomas have a similar presentation. While clinical and imaging features will help guide an appropriate work-up, any suspicious soft tissue mass

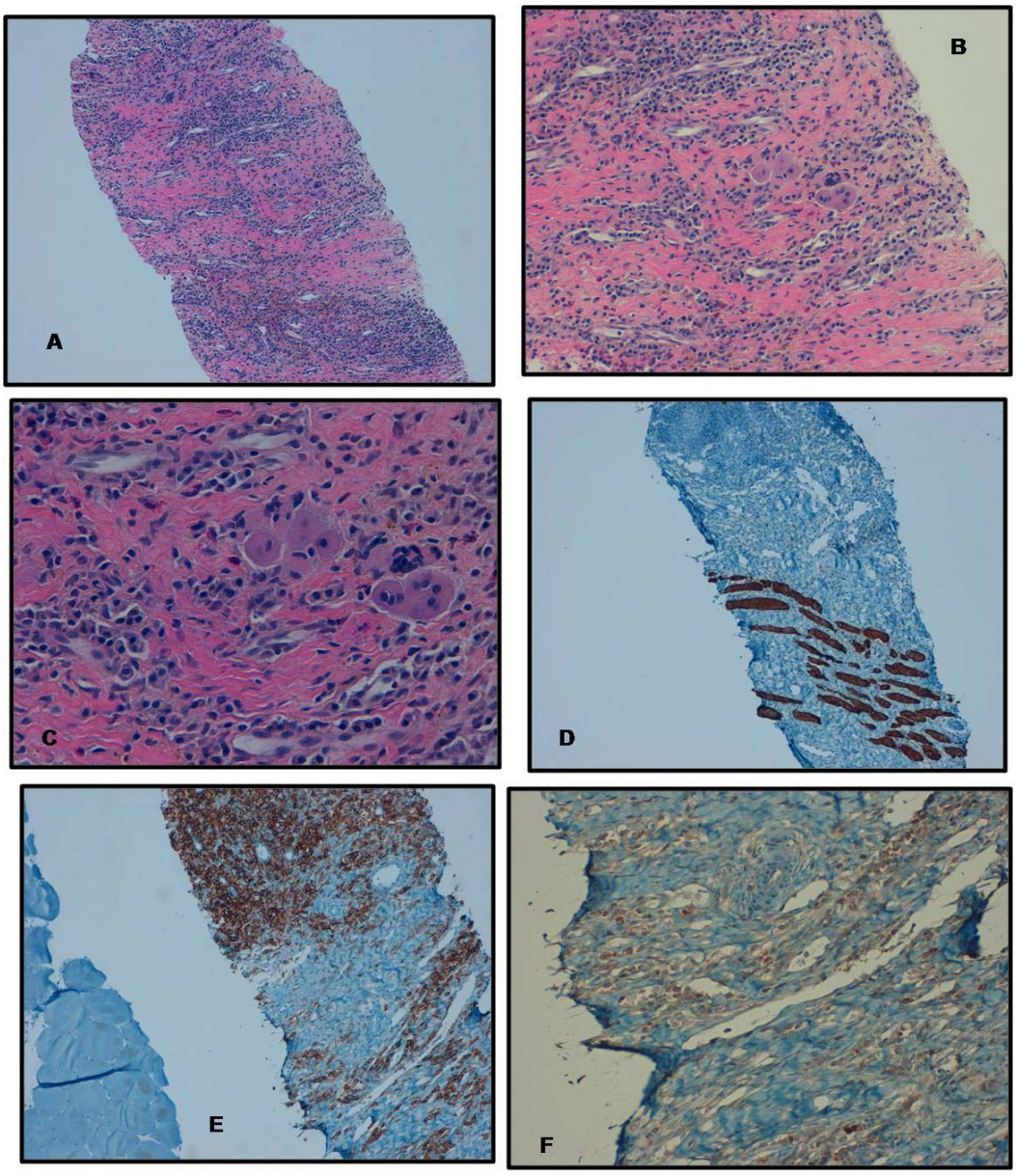

Figure 2. Histopathological Findings. (A)-(C) Sections of the lesion show a fibroinflammatory condition affecting the skeletal muscle (A, H & E, ×100. B, H & E, ×200. C, H & E, ×400). (D) Desmin stain ×100 positive for skeletal muscle. (E) CD 38 marker positive indicating plasma cell infiltration. (F) IgG4 stain positive revealed an increased IgG4 plasma cell count.

must be biopsied [1].

IgG4-RD is a novel lymphoproliferative syndrome of unknown etiology including a collection of disorders that share clinical and pathological characteristics. Several of the manifestations can occur in the same patient and comprise: Type 1 autoinmune pancreatitis and IgG4-related sclerosing cholangitis, inflammatory orbital pseudotumor, retroperitoneal fibrosis, thyroiditis, and other sclerosing or IgG4-related disorders as dacryoadenitis, sialadenitis, aortitis and periaortitis, interstitial pneumonitis, tubulointerstitial nephritis, hypertrophic pachymeningitis [2].

The principal symptoms and signs of IgG4-RD are a slow growing mass or diffuse enlargement of an organ. To our knowledge this is the first reported case of localized IgG4-related disease affecting skeletal muscle and presenting as a soft tissue tumor. IgG4-RD can also present with clinical findings related to affected organs with fibrotic or sclerotic changes [2].

Pathologically the disease is characterized by IgG4 positive plasma cells and lymphocytes tissue infiltration which may be accompanied by sclerosis. Elevated serum levels of IgG4 may be present, being an important aid in diagnosis, although they are not diagnostic [3].

The diagnosis of IgG4-RD is made with biopsy findings demonstrating the characteristic histopathology. Core-Needle Biopsy or Tru-Cut Biopsy is most often indicated for superficial or accessible deep extremity soft tissue tumors that are of sufficient size to needle placement (>3 cm) [1]. In our case this was the technique of biopsy indicated. A good therapeutic response to glucocorticoids is also characteristic of IgG4-RD, by symptomatic and organ function improvement or reductions in the size of masses, as it happened in our case [4,5].

Glucocorticoid therapy has been suggested as the initial treatment of IgG4-RD. Azathioprine, mycophenolate mofetil and rituximab have been used to treat resistant patients [5,6].

The natural history and prognosis of IgG4-related disease are not well described. Despite that good responsiveness to glucocorticoid treatment, relapses are common and organ dysfunction may arise from inflammatory and fibrotic changes. The possibility of increased risk of malignancy is not clear [5,6].

4. Conclusion

Despite its extreme rarity, IgG4-related disease may be presented as a soft tissue tumor affecting skeletal muscle. Biopsy may be the only diagnostic procedure. A good therapeutic response to glucocorticoids should be expected.

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